Rare disease

Multidetector CT of rare isolated partial anomalous pulmonary venous return

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Summary

A 16-year-old male was referred to our clinic for evaluation of pulmonary hypertension. An isolated anomalous pulmonary venous return of both left pulmonary veins to the innominate vein was diagnosed after echocardiographic and multidetector CT (MDCT) evaluation. Three-dimensional MDCT image reconstructions were used to better clarify this rare type of disease and adequate choice of surgical approach.

BACKGROUND

The following case report presents high-quality multi-detector CT (MDCT) images of a rare anatomical type of isolated partial anomalous pulmonary venous return (PAPVR).

We think it may be of interest to the readership of *BMJ Case Reports* journal due to the diagnostic investigation and the fact that it may help some clinicians not miss this, sometimes hard to diagnose, entity.

CASE PRESENTATION

A 16-year-old male was referred to our clinic for evaluation of pulmonary hypertension. The patient had complaints of exertional dyspnoea (NYHA class II), was overweight (104 kg vs 174 cm; 34.4 kg/m² body mass index – obese class I 30.1–34.9 kg/m²), but had no other relevant findings on physical examination.

INVESTIGATIONS

ECG was in sinus rhythm with incomplete right bundle branch block. Previously performed transthoracic echocardiogram with a bad acoustic window revealed moderate dilatation of right atrium and ventricle (RV diastolic diameter 51 mm; LV diastolic diameter 45 mm). Systolic pressure of the pulmonary artery (PAP) was estimated in 49 mm Hg (29 mm Hg RA/RV gradient plus 20 mm Hg due to lack of respiratory collapse of dilated inferior vena cava). Intracardiac shunts were excluded, namely atrial septal defect with transesophageal echocardiogram with agitated saline serum. Left pulmonary veins could not be visualised.

An MDCT was performed in our centre and PAPVR of both left pulmonary veins to the innominate vein was diagnosed (figures 1 and 2).

Preoperative invasive assessment was performed in the cath lab and an elevation of the average PAP (37 mm Hg)

Figure 1 Thoracic CT angiographic findings. (A) Axial contrast-enhanced image shows absence of drainage of the left pulmonary veins (white arrow heads) into the left atrium. (B) Right anterior oblique reconstruction showing drainage of the left pulmonary veins into innominate vein.

Figure 2 3D volume-rendered images in coronal view (A) and left sagittal view (B) illustrating drainage of the left pulmonary veins (PV) into the innominate vein (IV), through the vertical vein (VV).

alongside with volume overload of pulmonary circulation (Qp:Qs 2.1) were confirmed.

OUTCOME AND FOLLOW-UP

The patient underwent corrective surgery (anastomosis of the vertical vein into the left atrial appendage) and is now asymptomatic 1 year after.

DISCUSSION

The authors report a rare anatomical type of isolated PAPVR in a symptomatic patient with pulmonary hypertension. PAPVR occurs when one or more (but not all) pulmonary veins are abnormally connected to the right atrium, either directly or indirectly, via the superior or inferior vena cava. This entity is found in 0.4–0.7% in autopsy series of patients with congenital heart disease.¹ Even though involvement of the right pulmonary veins seems to be more common (from 2¹ to 10 times more frequent²), there have been some recent contradicting studies showing similar prevalence in a series accessed by MDCT.³ Simultaneous cardiac involvement (frequently atrial septal defect of sinus venosus type) is found in 90% of patients.⁴ PAPVR with intact septum (isolated PAPVR) is rare and commonly involves anomalous drainage of the right upper pulmonary vein into the superior vena cava. Drainage from the left lung into the innominate vein is extremely rare (only 3% of patients). In this patient, both left pulmonary veins were draining in the innominate vein through an ascending vessel, the vertical vein. Sometimes this type of PAPVR can also be diagnosed by an abnormal catheter position.⁵

The fact of having normal right side drainage and lack of an associated atrial septal defect may have lead to the mild clinical presentation and late adolescence diagnosis.

PAPVR can be easily missed in routine echocardiography if a high level of suspicion is not present and pulmonary

Learning points

- ▶ PAPVR can be easily missed in routine echocardiography if a high level of suspicion is not present and pulmonary veins are not routinely assessed (apical four-chamber view in transthoracic and at 45° in transesophageal echocardiogram).
- ▶ Careful assessment of the pulmonary veins is recommended in every patient with pulmonary hypertension and/or right heart enlargement.
- ▶ The drainage of both left pulmonary veins into the innominate vein through an ascending vessel, the vertical vein, may be observed as a rare variety of PAPVR.
- ▶ MDCT can non-invasively confirm diagnosis and provide precise anatomical details of the PAPVR in order to plan corrective surgery.
- ▶ Cardiac MRI may be a feasible alternative, or even first approach option, in the workup of potential extra-cardiac shunts due to radiation safety and functional evaluation advantages (namely Qp:Qs assessment).
- ▶ Qp:Qs assessment is of utmost importance for treatment decisions in PAPVR.
- ▶ Symptomatic patients with PAPVR with pulmonary circulation volume overload confirmed by Qp:Qs > 1.5 and dilatation of right heart cavities have a clear indication for surgical correction.

veins are not routinely assessed (apical four-chamber view in transthoracic and at 45° in transesophageal echocardiogram). Therefore, we recommend careful assessment of the pulmonary veins in every patient with pulmonary hypertension and/or right heart enlargement. MDCT can non-invasively confirm diagnosis and provided us with precise anatomical details of the PAPVR in order to plan corrective surgery. MDCT has also allowed us to exclude

the possibility of pulmonary embolism. Cardiac MRI may be a feasible alternative, or even first approach option, in the workup of potential extra-cardiac shunts due to radiation safety and functional evaluation advantages, namely the possibility of precise non-invasive Qp:Qs assessment using phase-velocity MRI to measure flow in the pulmonary artery and in the ascending aorta in millilitre per heart beat.⁶ Though, it was not an option for our patient due to limited access to this examination in our hospital.

This patient had a clear indication for surgery; he had a symptomatic PAPVR of the entire left lung with pulmonary circulation volume overload confirmed by Qp:Qs >1.5 and dilatation of right heart cavities. In asymptomatic patients without RV dilatation and Qp:Qs below 1.5, watchful waiting is advisable.⁷ Qp:Qs assessment is, therefore, of utmost importance for treatment decisions in PAPVR.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Zwetsch B**, Wicky S, Meuli R, *et al*. Three-dimensional image reconstruction of partial anomalous pulmonary venous return to the superior vena cava. *Chest* 1995;**108**:1743–5.
2. **Snellen HA**, van Ingen HC, Hoefsmit EC. Patterns of anomalous pulmonary venous drainage. *Circulation* 1968;**38**:45–63.
3. **Ho ML**, Bhalla S, Bierhals A, *et al*. MDCT of partial anomalous pulmonary venous return (PAPVR) in adults. *J Thorac Imaging* 2009;**24**:89–95.
4. **SHUMACKER HB Jr**, JUDD D. Partial anomalous pulmonary venous return with reference to drainage into the inferior vena cava and to an intact atrial septum. *J Cardiovasc Surg (Torino)* 1964;**5**:271–8.
5. **Klecker RJ**, Christoforidis AJ, Sinclair DS. Case 2. Vertical vein (partial anomalous pulmonary venous drainage). *AJR Am J Roentgenol* 2000;**175**:867; 869–70.
6. **Wang ZJ**, Reddy GP, Gotway MB, *et al*. Cardiovascular shunts: MR imaging evaluation. *Radiographics* 2003;**23**:S181–94.
7. **Vogel M**. Partial anomalous pulmonary venous connection and the scimitar syndrome. In: Gatzoulis MA, Webb GD, Daubeney PEF, eds. *Adult Congenital Heart Disease Diagnosis and Management*. Edinburgh: Churchill Livingstone 2003:205–210.

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